

**Ataxin3 (MJD) Antibody (N-term)**  
**Purified Rabbit Polyclonal Antibody (Pab)**  
**Catalog # AP2181a****Specification**

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**Ataxin3 (MJD) Antibody (N-term) - Product Information**

Application	IHC-P,E
Primary Accession	<a href="#">P54252</a>
Reactivity	Human
Host	Rabbit
Clonality	Polyclonal
Isotype	Rabbit IgG
Calculated MW	41250
Antigen Region	29-59

**Ataxin3 (MJD) Antibody (N-term) - Additional Information****Gene ID** 4287**Other Names**

Ataxin-3, Machado-Joseph disease protein 1, Spinocerebellar ataxia type 3 protein, ATXN3, ATX3, MJD, MJD1, SCA3

**Target/Specificity**

This Ataxin3 (MJD) antibody is generated from rabbits immunized with a KLH conjugated synthetic peptide between 29-59 amino acids from the N-terminal region of human Ataxin3 (MJD).

**Dilution**

IHC-P~~1:50~100

**Format**

Purified polyclonal antibody supplied in PBS with 0.09% (W/V) sodium azide. This antibody is prepared by Saturated Ammonium Sulfate (SAS) precipitation followed by dialysis against PBS.

**Storage**

Maintain refrigerated at 2-8°C for up to 2 weeks. For long term storage store at -20°C in small aliquots to prevent freeze-thaw cycles.

**Precautions**

Ataxin3 (MJD) Antibody (N-term) is for research use only and not for use in diagnostic or therapeutic procedures.

**Ataxin3 (MJD) Antibody (N-term) - Protein Information****Name** ATXN3 {ECO:0000303|PubMed:33157014, ECO:0000312|HGNC:HGNC:7106}**Function** Deubiquitinating enzyme involved in protein homeostasis maintenance, transcription, cytoskeleton regulation, myogenesis and degradation of misfolded chaperone substrates

(PubMed:[12297501](#), PubMed:[17696782](#), PubMed:[23625928](#), PubMed:[28445460](#), PubMed:[33157014](#), PubMed:[16118278](#)). Binds long polyubiquitin chains and trims them, while it has weak or no activity against chains of 4 or less ubiquitins (PubMed:[17696782](#)). Involved in degradation of misfolded chaperone substrates via its interaction with STUB1/CHIP: recruited to monoubiquitinated STUB1/CHIP, and restricts the length of ubiquitin chain attached to STUB1/CHIP substrates and preventing further chain extension (By similarity). Interacts with key regulators of transcription and represses transcription: acts as a histone-binding protein that regulates transcription (PubMed:[12297501](#)). Acts as a negative regulator of mTORC1 signaling in response to amino acid deprivation by mediating deubiquitination of RHEB, thereby promoting RHEB inactivation by the TSC-TBC complex (PubMed:[33157014](#)). Regulates autophagy via the deubiquitination of 'Lys-402' of BECN1 leading to the stabilization of BECN1 (PubMed:[28445460](#)).

#### Cellular Location

Nucleus matrix. Nucleus. Lysosome membrane; Peripheral membrane protein.  
Note=Predominantly nuclear, but not exclusively, inner nuclear matrix (PubMed:9580663).  
Recruited to lysosomal membrane in response to amino acid deprivation by the RagA/RRAGA-RagB/RRAGB complex (PubMed:33157014)

#### Tissue Location

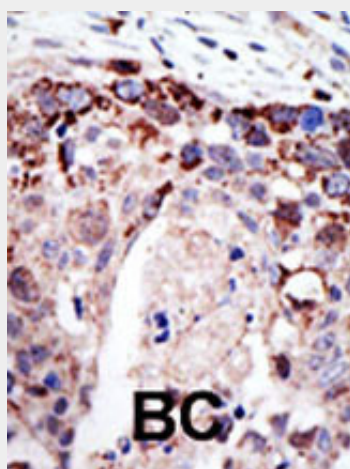
Ubiquitous.

### Ataxin3 (MJD) Antibody (N-term) - Protocols

Provided below are standard protocols that you may find useful for product applications.

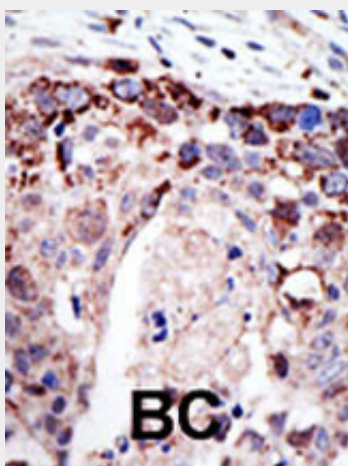
- [Western Blot](#)
- [Blocking Peptides](#)
- [Dot Blot](#)
- [Immunohistochemistry](#)
- [Immunofluorescence](#)
- [Immunoprecipitation](#)
- [Flow Cytometry](#)
- [Cell Culture](#)

### Ataxin3 (MJD) Antibody (N-term) - Images



Formalin-fixed and paraffin-embedded human cancer tissue reacted with the primary antibody, which was peroxidase-conjugated to the secondary antibody, followed by AEC staining. This data demonstrates the use of this antibody for immunohistochemistry; clinical relevance has not been

evaluated. BC = breast carcinoma; HC = hepatocarcinoma.



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#### **Ataxin3 (MJD) Antibody (N-term) - Background**

Machado-Joseph disease is an autosomal dominant neurologic disorder, and is now known to be the same as previously described spinocerebellar ataxia-3. MJD protein (Ataxin-3) contains (CAG) $n$  repeats in the coding region, and the expansion of these repeats from the normal 13-36 to 68-79 is the cause of Machado-Joseph disease. There is a negative correlation between the age of onset and CAG repeat numbers. This protein interacts with key regulators (CBP, p300 and PCAF) of transcription and represses transcription, and also acts as a histone-binding protein that regulates transcription. MJD is a deubiquitinating enzyme.

#### **Ataxin3 (MJD) Antibody (N-term) - References**

- Albrecht, M., et al., Eur. J. Biochem. 271(15):3155-3170 (2004).
- Michlewski, G., et al., J. Mol. Biol. 340(4):665-679 (2004).
- Li, Y., et al., Ann. Neurol. 56(1):124-129 (2004).
- Beuckmann, C.T., et al., J. Neurosci. 24(18):4469-4477 (2004).
- Berke, S.J., et al., J. Neurochem. 89(4):908-918 (2004).